

Solitary plasmacytoma of mandible: A rare entity

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Abstract

Plasmacytoma is a discrete, unifocal, monoclonal neoplastic proliferation of the plasma cells. It may present as one of the three distinct clinical entities: multiple myeloma (MM), solitary plasmacytoma of bone and extramedullary plasmacytoma. Solitary bone plasmacytoma accounts for 3% of all plasma cell neoplasms with approximately 50% of the cases transforming into MM. It is frequently seen in vertebrae and secondarily in long bones. Its presence in jaws is extremely rare and when it is seen, retromolar area, angle and ramus of the mandible are most common sites of occurrence. A comprehensive clinical, radiological, histological and immunohistochemical features of solitary plasmacytoma of the mandible in a 46-year-old female patient is reported.

Keywords: Mandible, plasma cells, plasmacytoma

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Received: 20.07.2018, **Accepted:** 23.01.2019

INTRODUCTION

Plasmacytomas are the diverse group of lymphoid neoplasms which are characterized by clonal neoplastic proliferation of terminally differentiated B-lymphocytes (plasma cells or myeloma cells).^[1] It was first mentioned by Unna in 1891 and described by Schridde in 1905. These are grouped under B-cell peripheral lymphomas according to the classification of the Revised European-American International Lymphoma Study Group.^[2] Plasma cell neoplasms account for approximately 2.6–3.3/100,000 population.^[2,3]

The three distinct clinical entities of plasma cell neoplasms include multiple myeloma (MM), solitary plasmacytoma of the bone (SPB) and extramedullary plasmacytoma (EMP). MM is the most common neoplasm accounting for 65% and also represents 1% of all malignancies.^[2] SPB

and EMP are less common, localized forms which further evolve into disseminated MM within months or years after the initial diagnosis.^[4]

The course of SPB is restricted to one medullary bone which accounts for 3%–7% of all plasma cell neoplasms.^[5] SPB primarily affects the axial skeleton, especially vertebrae and secondarily long bones. However, jaw involvement is very rare, and when it does, it shows a predilection for mandible than maxilla. Most of the cases reported in the literature are in the bone marrow-rich areas of the mandible that is in the body, angle, ramus and also retromolar trigone.^[6] SPB is considered to be a distinct entity or as first manifestation of subsequent MM.^[7]

A rare case of solitary plasmacytoma of the mandible was reported in a 46-year-old female patient suggesting the utmost importance of early diagnosis by the clinician.

Access this article online

Quick Response Code:



Website:

www.jomfp.in

DOI:

10.4103/jomfp.JOMFP_175_18

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How to cite this article: Chittamsetti S, Guttikonda VR, Sravya T, Manchikatla PK. Solitary plasmacytoma of mandible: A rare entity. J Oral Maxillofac Pathol 2019;23:136-9.

CASE REPORT

A 46-year-old female patient reported to the outpatient department of our hospital complaining of pain and swelling over the right middle-third region of the face for 1 month. Past dental history revealed extraction of 48 1 month back, after which she noticed a painful swelling which was initially peanut in size and gradually increased to attain the present size. Past medical history was noncontributory and general physical examination revealed no abnormality.

Extraoral examination revealed facial asymmetry on the right side of the face. On inspection, a solitary, diffuse swelling over the middle-third region of the face was noticed [Figure 1]. The overlying skin appeared to be normal. Swelling was roughly oval measuring approximately 5 cm × 6 cm, extending superoinferiorly from 1 cm below the infraorbital margin to lower border of the mandible and anteroposteriorly from 0.5 cm away from ala of the nose to tragus of the ear. On palpation, all the inspeitory findings were confirmed and the swelling was soft to firm in consistency. Single right submandibular lymph node was palpable, which is nontender and firm in consistency measuring approximately 1.5 cm × 1 cm.

Intraoral examination [Figure 2] revealed a well-defined solitary erythematous growth in the right retromolar region at the previous extraction site. The lesion was extending posteriorly from distal aspect of 47 to retromolar pad, measuring approximately 3 cm × 3 cm. On palpation, all the inspeitory findings were confirmed and growth was soft to firm in consistency with smooth surface and well-defined margins.

Orthopantomograph [Figure 3] revealed ill-defined radiolucency encompassing entire right side of the

mandible extending posteriorly from 47 to ramus, angle, coronoid and condylar process. Reconstructed computed tomography scan revealed large expansile lytic lesion with irregular borders on the right posterior mandible region [Figure 4a]. Appreciable erosive changes of alveolar surface posterior to 47 were noted. Medial aspect of the ramus was completely resorbed [Figure 4b]. Based on the above clinical and radiological findings, a provisional diagnosis of intraosseous carcinoma of the right retromolar area was given. However, central giant cell lesion and osteosarcoma were considered in the differential diagnosis.

An incisional biopsy was performed and sent for histopathological examination. Histopathology revealed solid sheets of densely packed, uniformly appearing round-to-ovoid cells with eccentrically placed nucleus resembling plasma cells [Figure 5a]. These plasma cells are scattered over background of numerous lymphocytes with relatively sparse stroma. Anaplastic cells with hyperchromatic nucleus and occasional binucleated forms [Figure 5b] suggesting malignancy were also observed. Based on the above findings, a histopathological diagnosis of plasmacytoid non-Hodgkin's lymphoma was given. Differential diagnosis of plasmacytoma and large-cell lymphoma of immunoblastic type was considered.

To confirm the diagnosis, laboratory investigations such as routine blood investigations were performed which revealed elevated serum-free lambda light chains to 325.9 mg/L where the normal levels range from 5.71 to 26.30 mg/L. Based on laboratory findings, the final diagnosis of solitary plasmacytoma of the mandible was given which was then confirmed by immunohistochemistry which revealed strong positivity for CD138 [Figure 6a], MUM1 [Figure 6b], variable cell



Figure 1: Extraoral examination revealing facial asymmetry on the right side of the face



Figure 2: Intraoral examination revealing a well-defined solitary erythematous growth in the right retromolar region



Figure 3: Orthopantomograph revealing ill-defined radiolucency encompassing the entire right side of the mandible

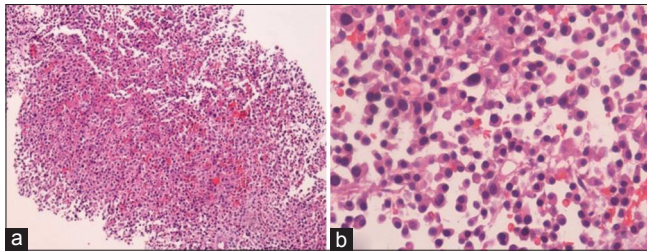


Figure 5: Photomicrograph showing solid sheets of plasma cells (a) and also anaplastic cells with occasional binucleated forms (b)

membrane positivity for CD45 [Figure 6c] and negativity for CD20 [Figure 6d].

DISCUSSION

SPB is an immunoproliferative monoclonal disease which rarely affects the jaws and manifests as a single osteolytic lesion. The etiology of SPB remains uncertain, but several hypotheses were proposed which implicated the role of radiation, chemical exposure, viruses and genetic factors. Cytogenetic studies revealed loss in chromosome 13, 1p, 14q and gain in 19p, 9q, 1q, and interleukin-6 is considered as principal growth factor in pathogenesis.^[5]

SPB occurs in slightly younger age group than MM with an average age of diagnosis being 55 years^[8] and is more prevalent in males than females with ratio being 2:1.^[3,5] But in contrast to the literature, the present case was reported at slightly earlier age of 46 years in a female patient.

Although SPB is uncommon in head and neck, only 4.4% of the cases occur in bone marrow-rich areas of posterior mandible.^[9] The present case was reported in posterior mandible consistent with a study conducted by Pisano *et al.* in which, among 13 cases of SPB, 9 lesions were reported posterior to premolar area.^[8]

Most common clinical symptoms of SPB are pain in jaws, paresthesia, mobility, hemorrhage, swelling in hard and soft tissues and pathological fractures.^[5,8] The present case was in harmony with the literature as the swelling was associated with pain and was erythematous in color.



Figure 4: Reconstructed computed tomography scan revealing large expansile lytic on the right posterior mandible region (a) Completely resorbed Medial aspect of the ramus with alveolar erosion(b)

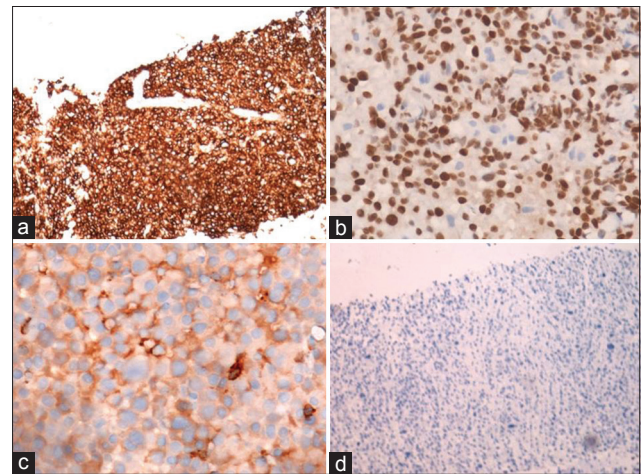


Figure 6: Immunohistochemistry showing strong positivity for CD138 (a), MUM1 (b), variable cell membrane positivity for CD45 (c) and negativity for CD20 (d)

Malignant plasma cells produce cytokines and release osteoclast-activating factor which stimulates osteoclasts to resorb the bone; therefore, on radiographic examination, they present as well-defined osteolytic lesions with unilocular or multilocular radiolucency.^[5,6] According to Lae *et al.*,^[10] there are three radiographic patterns in SPB, which include multilocular soap-bubble lesions, unilocular radiolucency with cystic appearance and ill-defined destructive bone resorption. In the present case, an ill-defined radiolucency with extensive cortical destruction of entire posterior aspect of the mandible was noticed.

Histopathologically, SPB presents as clusters or sheets of atypical plasma cells with varying degree of differentiation and sparse stroma. Plasma cells are characterized by abundant cytoplasm with eccentrically placed nucleus which may often show chromatin clumps typically arranged in cartwheel or clock-face pattern. Occasionally binucleated cells also noticed.^[4,5] The present case was also in accordance with the previous findings. Sometimes, cells may also show paranuclear, pale staining area called as “hof.” Giant cell formation, amyloid deposition and myxoid change and plasma cell inclusion bodies can be noticed in few cases.^[4,5]

Table 1: Specific diagnostic criteria for solitary Plasmacytoma of Bone

Criteria
Isolated tumor composed of malignant plasma cells
Absence of other lesions on skeletal radiographic survey
Absence of plasmacytosis in the bone marrow
Absence of anemia, hypercalcemia or renal involvement

Sukpanichnant *et al.*^[11] categorized plasmacytomas as mild, moderate and severe dysplasia based on the degree of plasma cell differentiation. The present case was considered under mild dysplasia as there are more mature plasma cells.

The neoplastic process is secretory in about 99% of cases producing monoclonal light chain or heavy chain immunoglobulins which are identified in serum or urine.^[10] In concordant with the literature, the present case shows elevated serum lambda light chains to greater amount than normal. To diagnose a case of SPB and to rule out MM, careful clinical investigations must be done which include skeletal radiological survey, bone marrow aspiration, blood cell count, determination of calcium levels and study of renal function.^[10] Specific diagnostic criteria for SPB were given by Bataille and Sany^[12] [Table 1]. The present case satisfies all the mentioned criteria. In addition to laboratory investigations, monoclonal restriction to either lambda or kappa light chains is an essential approach to evaluate the suspected plasmacytoma.

Plasmacytoma has to be differentiated from reactive inflammatory lesions such as plasma cell gingivitis, non-Hodgkins lymphoma and malignant melanoma which presents a diagnostic challenge to clinicians.^[5]

Presence of nonplasmacytic neoplastic component, IgM expression and positivity for pan B-cell surface markers such as CD19, CD20 and CD79a favors diagnosis of lymphomas. Immunohistochemical studies are used to rule out malignant melanoma which shows positive expression for S100 and HMG-45 markers, whereas plasmacytoma shows negative expression of these markers.^[4]

The course of SPB is relatively benign and the prognosis was better than MM. However, if recurrence is present showing tendency toward MM, the prognosis is worse. Survival rate is 50%–80% at 10-year duration and if recurrence is noticed, it drops down to 16%.^[3]

CONCLUSION

Plasma cell tumors of head and neck in the absence of myelomatosis are rare; therefore, dental surgeons

necessitate the knowledge of oral manifestations of SPB which is important for early diagnosis of the disease to render optimal treatment. As there is definite risk of transformation of SPB into subsequent disseminated disease, continued follow-up is recommended.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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